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Original Research Paper

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CHOLELITHIASIS, PANCREATIC DIVISUM IN POLYCYSTIC KIDNEY DISEASE :A RARE CONCURRENCE

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ABSTRACT Autosomal-dominant polycystic kidney disease (ADPKD) is the most common inherited kidney disease. It is characterized by multiple cysts of variable size diffusely scattered throughout the renal cortex and medulla [1]. It is a systemic disorder leading to chronic renal failure and hypertension. Autosomal dominant polycystic kidney disease (ADPKD) is an inherited disorder associated with multiple cyst formation in the different organs. Extrarenal cysts are mainly found in liver, but may involve pancreas, spleen and brain but with lesser frequency. Affected individuals have a relatively high rate of cardiac valvular abnormalities, especially mitral valve prolapse, as well as intracranial, aortic or coronary aneurysms, abdominal and inguinal hernias and colonic diverticulosis [1] We report a case with both pancreas divisum and cholelithiasis in patient of ADPKD.A 34-year-old gentleman with ADPKD presented with pain upper abdomen associated with vomiting and fever. Imaging studies revealed multiple renal and hepatic cysts, cholelithiasis, pancreatitis and acute fluid collection.Pancreatitis is common cause of abdominal pain in patients with ADPKD. Pancreas divisum may be a predisposing factor for acute pancreatitis in ADPKD.

KEYWORDS : Autosomal Dominant Polycystic Kidney Disease (adpkd), Extrarenal Cysts, Pancreas Divisum

CASE PRESENTATION

A 34-year-old man was admitted to the Department Of Minimal Access Surgery with vomiting, fever and pain in the upper abdomen radiating to the back, since 10 days. Five days before, the patient had been admitted to another center with the diagnosis of pancreatitis but did not improve. Patient was diagnosed with ADPKD, five years back. He was also hypertensive since then. He denied using alcohol or any other drugs. There was no history of either trauma or surgery. Physical examination was apparently unremarkable. Vital were normal. There was tenderness in the epigastrium.

PHYSICAL AND EXAMINATION :

During the hospitalization laboratory tests were as follows: white blood cell count, 6450/mm3 ; hemoglobin, 12.3 g/dl; platelets, 154,000/mm3 ; urea, 11 mg/dl (5–23); creatinine, 1.18 mg/dl (0.6–1.3); alanine aminotransferase, 16 U/l (0–40); aspartate aminotransferase, 21 U/l (0–40); alkaline phosp hat ase, 141 U/l (38–155); gamma-glutamyltransferase, 46 U/l (0–55); total bilirubin, 0.46 mg/dl (0.1–1.0); direct bilirubin, 0.12 mg/dl (0.1–0.2); albumin, 4.0 g/dl (3.5–5); amylase, 284 U/l (5–100).cholesterol, 122 mg/dl (120–190); triglycerides, 132 mg/dl (50–200); INR, 1.1; HBs Ag (–); anti HBcIgM (–); anti HCV (–); anti-HIV (–)Abdominal ultrasonography revealed multiple cystic lesions in liver and both kidneys, cholelithiasis and bulky pancreas. MRCP with MRI upper abdomen confirmed multiple cyst in both kidneys and liver, cholelithiasis, pancreatic divisum, pancreatitis with acute fluid collection.

OUTCOME AND FOLLOW UP

Hence, diagnosis of acute biliary pancreatitis was estab lished. Patient was resuscitated and then underwent laparoscopic cholecystectomy in same admission. Surgery was uneventful and patient improved symptomatically after surgery.

DISCUSSION

Autosomal dominant polycystic kidney disease, the most frequent inherited polycystic disease, is a systemic disorder characterised by the development of numerous and bilateral kidney cysts leading to chronic renal failure and hypertention. Extrarenal cysts are located mainly in the liver but also in various organs including the pancreas.About 60% of ADPKD patients have abdominal or back pain which is usually related to effects of renal or liver cysts . Acute severe pain in ADPKD is usually due to hemorrhage, infection of the cysts or nephrolithiasis .[2-3]

Malka D et al reported chronic obstructive pancreatitis due to a pancreatic cyst in a patient with autosomal dominant polycystic kidney disease .Abdominal transparietal and endoscopic ultrasonography, computed tomography, and endoscopic retrograde cholangiopancreatography showed a cystic lesion in the body of the pancreas associated with upstream dilatation of the main pancreatic duct. Intraop erative ultrasonography before and after cyst fluid aspiration, and pancreatography and pathological examination of the resected distal pancreas confirmed that both main pancreatic duct enlargement and chronic pancreatitis were caused by a benign cyst.It was concluded that chronic obstructive pancreatitis should be added to the extrarenal complications of autosomal dominant polycystic kidney disease .[4]

Ultrasonographic study of pancreatic cysts in autosomal dominant polycystic kidney disease was done by Torra R et al . They studied 173 ADPKD patients and 160 non-affected family members and found a prevalence of pancreatic cysts, of 9% in ADPKD patients over 30 years of age. The presence of pancreatic cysts was related to the increasing age, to the female sex and to the type of ADPKD, found exclusively in PKD1 patients. No complications related to pancreatic cysts were recorded. They concluded that pancreatic cysts are an unusual feature of ADPKD and do not appear to contribute to morbidity or mortality .[5]

In a similar case like ours , Başar Ö et al saw recurrent pancreatitis in a patient with autosomal-dominant polycystic kidney disease. A 63-year-old man was evaluated for the etiology of recurrent pancreatitis and chronic renal failure. Multiple cysts of kidneys, liver, and pancreas and pancreas divisum was diagnosed. Pancreas divisum may be a predisposing factor for acute pancreatitis in these patients.[6]

Yazdanpanah K reporteda case of recurrent acute panc reatitis and cholangitis in a patient with autosomal dominant polycystic kidney disease. A 38-year-old man with ADPKD was presented with six episodes of acute pancreatitis and two episodes of cholangitis in a period of 12 months. Various ima ging studies revealed multiple renal, hepatic and pancreatic cysts, mild ectasia of pancreatic duct, dilation of biliary

system and absence of biliary stone. He was managed with conservative treatment for each attack. ADPKD should be considered as a potential risk factor for recurrent acute and/or chronic pancreatitis and cholangitis .[7]

Kim JA et al studied pancreatic cysts in autosomal dominant polycystic kidney disease, prevalence and association with PKD2 gene mutations. It was seen that patients with ADPKD were significantly more likely than control subjects to have at least one pancreatic cyst . In a univariate analysis, pancreatic cysts were more prevalent in patients with ADPKD with mutations in *PKD2* than in *PKD1* after adjusting for age, race, sex, estimated glomerular filtration rate, liver volume, and total kidney volume.[8]

However, in our patient, clinical and laboratory findings were suggestive of biliary pancreatitis. Although amylase levels can rise in many conditions, this patient's extremely high amylase along with an increased lipase suggested acute pancreatitis. The other causes of pancreatitis were ruled out: there was no history of alcohol, drug and herbal medicine, surgery, trauma or hereditary pancreatitis.We also ruled out hyperlipidemia and hypercalcemia. In patients with ADPKD, cysts occur in the liver in about 30% of cases and in pancreas in about 10% of cases . Pancreas divisum is most common congenital anomaly of the pancreas resulting from failure of fusion of the embryonic dorsal and ventral pancreatic ducts. There is controversy about a direct relationship between pancreas divisum and pancreatitis . Pancreas divisum may be associated with other biliary system anomalies such as long common channel, choledochal cysts and santorinicele, which may also predispose patients to pancreatitis . To our knowledge, renal and pancreatic cystic disease together with pancreas divisum has been reported very rarely in the literature.

CONCLUSION

Pancreatic pathologies should be included in the differential diagnosis of abdominal pain in patients with ADPKD. Pancreas divisum may be a predisposing factor for acute pancreatitis in these patients

CONFLICT OF INTERESTS

There are no financial or funding grants for this article. There is no conflict of interest to be disclosed.

ETHICAL APPROVAL It is a case report not a research study.

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AUTHOR'S CONTRIBUTION

All authors have equally contributed in collecting the data and preparing this article and are in agreement for publishing it.

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